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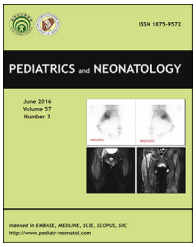

CASE REPORT

Scimitar Syndrome and H-type Tracheo-esophageal Fistula in a Newborn Infant

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Scimitar syndrome is a rare congenital anomaly characterized by partial anomalous pulmonary venous drainage of the right lung to the inferior vena cava (IVC) creating a tubular opacity paralleling the right cardiac border on chest radiography which resembles a curved Turkish sword or scimitar. Associated pulmonary and vascular anomalies have been reported in cases of Scimitar syndrome, most commonly hypoplasia of right lung, dextroposition of the heart, hypoplasia of the right pulmonary artery, and aberrant arterial supply from the descending aorta to the affected lobe of the right lung. To the best of our knowledge, this is the first case of Scimitar syndrome with an H-type tracheoesophageal fistula that has ever been reported.

1. Case Report

A male term newborn infant was born via vaginal delivery to a 22-year-old female. At birth, the infant was vigorous with spontaneous respirations and normal Apgar Scores. Besides a two-vessel umbilical cord, the infant appeared to be normal and was roomed in with the mother. At a few hours of life, a choking episode with discoloration was noted during the first bottle feed. Repeat examination revealed more prominent heart sounds on the right side of the chest. A chest x-ray revealed a cardiac silhouette that occupied right hemithorax (Figure 1). The infant was transferred to our institution for further evaluation.

Upon arrival, the infant appeared well and was breathing comfortably on room air. An echocardiogram

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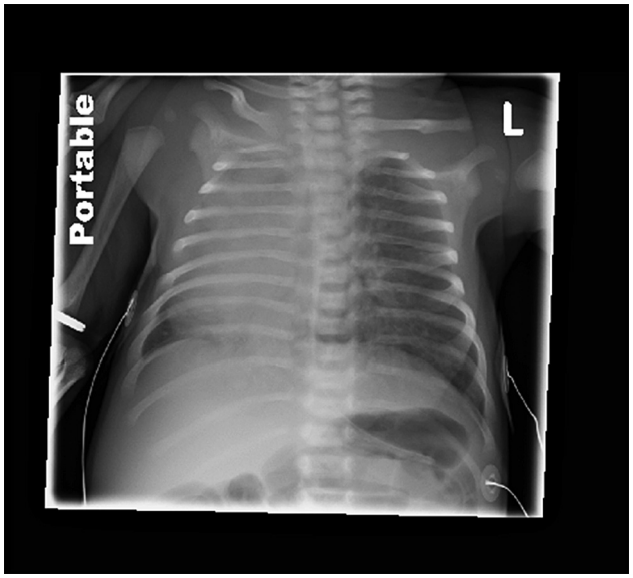

Figure 1 Chest radiography demonstrated the dextroposition of the cardiac silhouette occupies almost entire right chest.

demonstrated situs solitus, dextrocardia, concordant atrioventricular and ventricular great arterial connections, with normal drainage of the left-side pulmonary veins to the left atrium, but the connection of the right-sided pulmonary veins was not defined. In addition, there was evidence of suprasystemic pulmonary hypertension with bidirectional shunting through a 2-mm patent ductus arteriosus (PDA). The infant was started on oxygen via nasal cannula for pulmonary hypertension. He continued having persistent tachypnea despite normal oxygen saturation. A repeat echocardiogram demonstrated partial anomalous connection of the right pulmonary veins to the inferior vena cava with mild obstruction of the Scimitar vein at the diaphragm and persistent pulmonary hypertension (Figure 2E and F). He was started on furosemide and was weaned to room air successfully. Magnetic resonance angiography (MRA) confirmed the diagnosis of Scimitar syndrome: anomalous pulmonary venous drainage of all right-sided pulmonary veins to a vertical, scimitar-like vein that crossed the diaphragm and emptied into the IVC; there was normal left-sided pulmonary venous drainage, hypoplasia of the right lung, and a very small (2 mm) systemic arterial collateral arising from the abdominal aorta supplying the right lower lobe (Figure 2A—D). Repeat echocardiography demonstrated improvement of his pulmonary artery hypertension and spontaneous closure of ductus arteriosus. A decision was made to continue medical management with furosemide, maintain close follow-up and perform surgical repair of the partial anomalous pulmonary venous return at a later date.

Initially, a nasogastric tube was placed without difficulty and he tolerated the tube feeds. As his tachypnea resolved and he was able to maintain his saturation in room air, oral feeding was reintroduced and the episodes of choking and gasping for air during bottle feeding were noted. At that point his feeding difficulty was felt to be unrelated to his cardiac defect. Further evaluation by speech therapy noted the abrupt choking and aspiration after taking only a small volume of formula. A swallowing barium esophagogram was then obtained and demonstrated an H-type tracheoesophageal fistula (Figure 3).

Bronchoscopy confirmed the presence of the fistula in the membranous portion of the cervical trachea. Ligation and division of the H-type tracheoesophageal fistula was performed through a right-side incision. He recovered well and was kept NPO (*nil per os*) for 4 days. A repeat barium esophagogram demonstrated no esophageal leakage. He

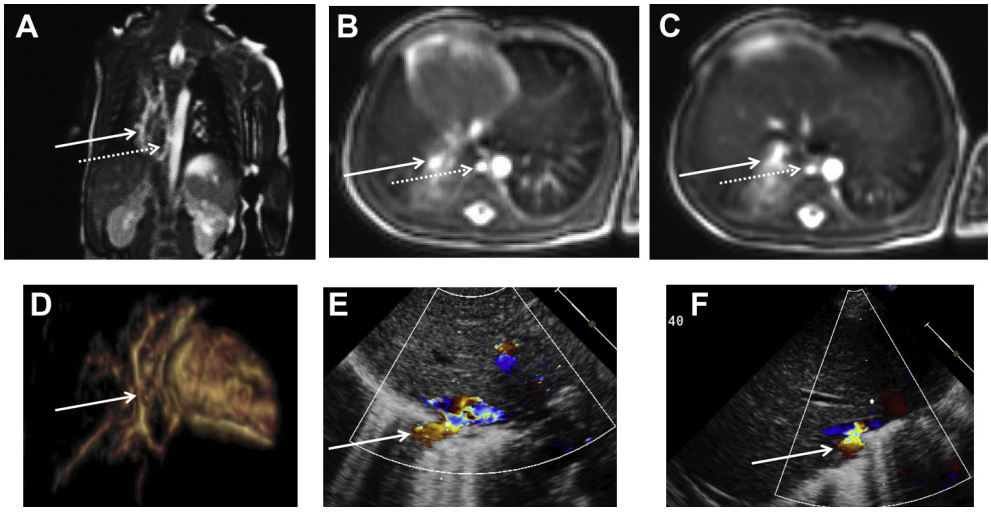

Figure 2 (A—C) Steady-state free precession magnetic resonance images. (A) Coronal view demonstrates the scimitar vein (white arrow) descending to the level of the diaphragm, and the azygous vein (dotted arrow). (B,C) Axial images reveal the scimitar vein crossing the diaphragm and inserting into the infradiaphragmatic, intrahepatic portion of the inferior vena cava (IVC); the azygous vein is also seen. (D) Three-dimensional reconstruction of gadolinium-enhanced magnetic resonance angiography (MRA) from a right posterior view displays the Scimitar vein. (E,F) Modified axial color Doppler echocardiographic views demonstrate similar findings to MRI views in B and C.

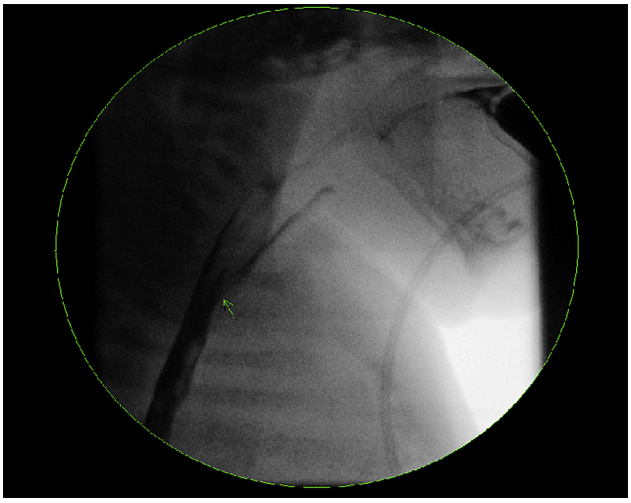

Figure 3 Esophagogram demonstrates the contrast media enter to the tracheal (arrow).

was gradually advanced to full feed. He was discharged in stable condition with a plan of close follow-up with pediatric cardiology and pediatric cardiothoracic surgery for continued evaluation and treatment for his Scimitar syndrome.

2. Discussion

Scimitar syndrome is a rare form of partial anomalous pulmonary venous return (PAPVR). PAPVR by itself is relatively uncommon and constitutes 0.5—1% of all congenital heart disease.[1] Only 3—6% of patients with PAPVR have Scimitar syndrome.[2] The true incidence of Scimitar syndrome is unknown since many of the patients are asymptomatic but an approximate incidence of 1 to 3 per 100,000 live births has been reported.[3] In this syndrome, there is partial or total anomalous venous drainage of the right lung into the inferior vena cava. The descending anomalous vein creates the curving soft tissue shadow along the right cardiac border on chest radiography which resembles the short curved Turkish sword or "scimitar"; hence, it was named "Scimitar syndrome".[2] It has also been termed "hypogenic lung syndrome" or the "venolobar syndrome" in the previous literature. According to the original definition, the syndrome is commonly associated with hypoplasia of the right lung, pulmonary sequestration, a persistent left superior vena cava, and dextroposition of the heart.[4] Scimitar syndrome is almost exclusively a right-sided anomaly; there are only a few case reports involving the left lung and left pulmonary veins.[5—7]

Associated congenital cardiovascular anomalies are seen in 75% of neonates and 36% of children with Scimitar syndrome. These include atrial septum defect (ASD), ventricular septum defect (VSD), coarctation of aorta, abnormalities of aortic arch, and abnormal relationship of the pulmonary arteries and bronchi.[8] Common non-cardiovascular anomalies include horseshoe lungs, small right hemithorax with diaphragmatic eventration, vertebral anomalies, scoliosis, hepatic herniation, absent inferior vena cava, and meningomyelocele.[9—12] One case with Scimitar syndrome and VATER association (vertebral defects, imperforate anus, trachea-esophageal fistula, and radial and renal dysplasia) has been reported, but detail of TE-fistula was not described.[13] This presenting case, to the best of our knowledge, is the first H-type tracheoesophageal fistula in a patient with Scimitar syndrome.

Patients with Scimitar syndrome have varied clinical presentations. The diagnosis in many adult patients is made incidentally. The common clinical presentation in infancy is respiratory distress and tachypnea due to associated pulmonary hypoplasia, pulmonary overcirculation, and/or pulmonary hypertension. Huddletson et al reviewed 12 cases of Scimitar syndrome in infancy; tachypnea was the most common clinical presentation with an average age of presentation of 6 weeks after birth.[14] The triad of respiratory distress, right lung hypoplasia and dextroposition of the heart should raise the suspicion of this syndrome.[8] Wheezing and recurrent pulmonary infections are the common clinical characteristics in pediatric patients with Scimitar syndrome.[4,14,15] Retrospectively, our patient presented with feeding difficulties from the beginning due to the H-type TE fistula, but his clinical presentation was overshadowed by other signs and symptoms related to his Scimitar syndrome: pulmonary hypertension, right lung hypoplasia, and congestive heart failure. The Scimitar sign on chest radiography may not be present in infants with Scimitar syndrome, as in our case, due to significant dextroposition of the heart and/or small size of the Scimitar vein as previously described. In our case, the diagnosis of Scimitar syndrome was suspected by echocardiography and confirmed by MRA at 1 week of life. In the largest reported series of 32 patients with Scimitar syndrome by Najm et al,[13] the age of presentation was a strong predictor of the clinical course, with patients presenting in the 1st year of life being more likely to have congestive heart failure, pulmonary hypertension, and associated cardiovascular abnormalities.

Right lung hypoplasia, mild pulmonary venous obstruction, pulmonary over-circulation, and the presence of a small systemic artery supplying the right lower lobe may have contributed to the pulmonary hypertension in our patient. The H-type TE fistula with recurrent aspiration/micro-aspiration may be another contributing factor. Diuresis and subsequent surgical repair of the H-type TE fistula resulted in reducing pulmonary arterial pressure allowing the oxygen to be weaned off in our patient. Signs and symptoms of right-sided volume overload, worsening of pulmonary venous obstruction at the diaphragm, and the persistence and degree of pulmonary hypertension will determine the need for the surgical repair of the Scimitar syndrome. It would be ideal to delay the surgical repair beyond the first year of life, if clinically feasible, since repair of the anomalous veins in the first year of life is frequently associated with higher mortality and higher residual pulmonary venous obstruction.[13]

References

1. Halasz NA, Halloran KH, Liebow AA. Bronchial and arterial anomalies with drainage of the right lung into the inferior vena cava. *Circulation* 1956;**14**:826—46.

2. Neill CA, Ferencz C, Sabiston DC, Sheldon H. The familial occurrence of hypoplastic right lung with systemic arterial supply and venous drainage, "Scimitar syndrome". *Bull Johns Hopkins Hosp* 1960;**107**:1—21.
3. Mathey J, Galey JJ, Logeais Y, Santoro E, Vanetti A, Maurel A, et al. Anomalous pulmonary venous return into inferio vena cava and associated bronchovascular anomalies (the scimitar syndrome). *Thorax* 1968;**23**:398—407.
4. Kramer U, Dürnberger V, Fenchel M, Stauder N, Claussen CD, Miller S. Scimitar syndrome: morphological diagnosis and assessment of hemodynamic significance by magnetic resonance imaging. *Eur Radiol* 2003;**13**:L147—50.
5. D'Cruz IA, Arcilla RA. Anomalous venous drainage of the left lung into the inferior vena cava: a case report. *Am Heart J* 1964;**67**:539—44.
6. Mardini MK, Sakati NA, Lewall DB, Christie R, Nyhan WL. Scimitar syndrome. *Clin Pediatr (Phila)* 1982;**21**:350—4.
7. Rutledge JM, Hiatt PW, Wesley Vick G 3rd, Grifka RG. A sword for the left hand: an unusual case of left-sided scimitar syndrome. *Pediatr Cardiol* 2001;**22**:350—2.
8. Midyat L, Demir E, Aşkin M, Gülen F, Ulger Z, Tanaç R, et al. Eponym. Scimitar syndrome. *Eur J Pediatr* 2010;**169**:1171—7.
9. Dupuis C, Charaf LA, Brevière GM, Abou P. "Infantile" form of the scimitar syndrome with pulmonary hypertension. *Am J Cardiol* 1993;**71**:1326—30.
10. Gao YA, Burrows PE, Benson LN, Robinovitch M, Freedom RM. Scimitar syndrome in infancy. *J Am Coll Cardiol* 1993;**22**:873—82.
11. Canter CE, Martin TC, Spray TL, Weldon CS, Strauss AW. Scimitar syndrome in childhood. *Am J Cardiol* 1986;**58**:652—4.
12. Woodring JH, Howard TA, Kanga JF. Congenital pulmonary venolobar syndrome revisited. *Radiographics* 1994;**14**:349—69.
13. Najm HK, Williams WG, Coles JG, Rebeyka IM, Freedom RM. Scimitar syndrome: twenty years' experience and results of repair. *J Thorac Cardiovasc Surg* 1996;**112**:1161—9.
14. Huddleston CB, Exil V, Canter CE, Mendeloff EN. Scimitar syndrome presenting in infancy. *Ann Thorac Surg* 1999;**67**:154—60.
15. Suri D, Sodhi KS, Muralidharan J, Manoj R, Singhi S. Scimitar syndrome: an uncommon cause of wheezing. *Pediatr Emerg Care* 2008;**24**:164—6.